ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 12 | December - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

AGGRESSIVE ANGIOMYXOMA AN UNUSUAL VULVAL GROWTH – A CASE REPORT

Obstetrics & Gynaecology

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ABSTRACT

Angiomyxoma is a rare soft tissue tumour which is usually benign in nature. Two types of angiomyxoma have been identified- 1. Superficial. 2. Aggressive. It is a tumour composed of mixture of blood vessels in a myxoid or collagenous stroma. They usually arise from the pelvic or the perineal region. This tumour has a tendency to recur. Diagnosis of this tumour is mainly on the basis of histopathological examination. This is a case report of Aggressive Angiomyxoma occurring in a 25 year old, nulligravida patient who was treated surgically

KEYWORDS

INTRODUCTION

Angiomyxomas are rare soft tissue tumors, which are locally invasive and is most commonly seen over the perineum and vulvovaginal region. It affects women of reproductive age group and are usually distinctive from other vulval masses. These tumors present as a polyp or a pedunculated mass and have a high recurrence rate [1]. Recurrence may manifest many years (often decades) after the initial excision. Distant metastases and death due to aggressive angiomyxoma are exceedingly rare; two cases with such an outcome have been reported [2].

These tumors develop from a type from a type of cell called as myxoid cells which are found in the connective tissues.

We hereby recount a similar case of Vulval Angiomyxoma, treated surgically by local excision.

Case Report

Here is a case of a 25 year old, nulligravida, recently married, who presented to our outpatient department with complaints of a mass protruding from her right labia majora since a few months. The mass was initially small, but showed a progressive increase in size over 2 months. It was painless in nature with a foul smell. She has no other complaints.

On Local examination, a large pedunculated mass of 6 x 5 cm was seen jutting out from the right labia majora, which appeared to be fleshy, with areas of ulceration over its inferior and posterior aspect. It was painless on palpation with no evidence of any bleeding or discharge. Inguinal lymph nodes were not palpable.

On per speculum examination, she had a foul-smelling vaginal discharge with a small Nabothian cyst over the posterior lip of the cervix. Abdominal and vaginal examination were unremarkable.

A local Ultrasonography was done which was suggestive of a pedunculated polypoidal mass arising from labia majora with significant vascularity in the pedicle. There was no evidence of significant inguinal lymphadenopathy.

Patient was admitted and started on injectable antibiotics. The vulval mass was resected by clamping the vascular pedicle, cutting the growth and ligating it securely and confirming hemostasis. The specimen was sent for histopathological examination.

On cut section, a homogenous, greyish white glistening surface was seen. Microscopic examination showed a tissue lined by stratified squamous epithelium with dermis showing a diffuse tumor comprising of bland spindled to stellate cells and variable sized blood vessels in dense myxoid to collagenous stroma. Individual cells have round to oval bland nuclei with eosinophilic cytoplasm. Also seen are entrapped muscle fibers in collagenized stroma. No evidence of mitosis/ necrosis/ atypia/ malignancy seen. The final impression was that of an Angiomyxoma.

The patient made an uneventful recovery and was discharged by third day. Patient remains asymptomatic on follow up till date.

Fig 1: Gross picture of the mass with vascular pedicle

Fig 2: Posterior surface of the mass showing an ulcer

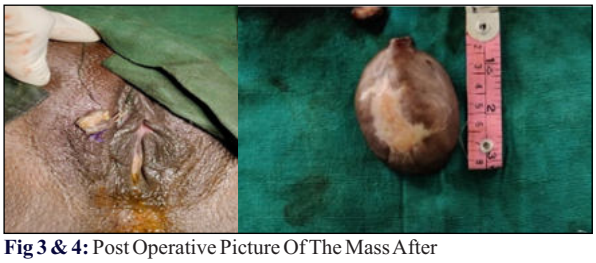

Fig 3 & 4: Post Operative Picture Of The Mass After

HISTOPATHOLOGY-

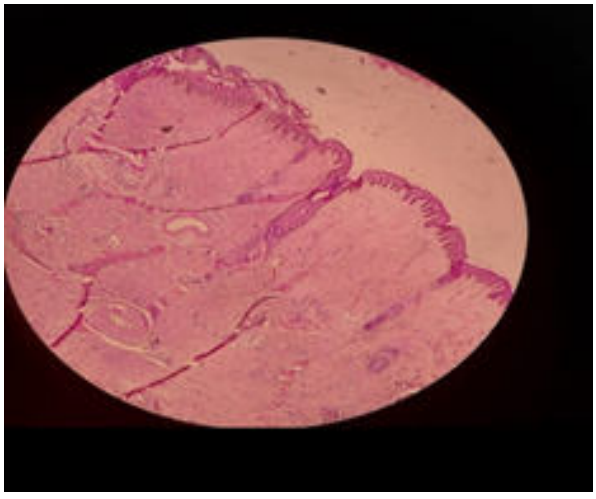

Fig 5: Cross section of the slide showing epithelial lining and stroma. The stroma is hypocellular and tumor is unencapsulated and locally infiltrative. Stroma shows myxoid changes with scattered collagen fibers

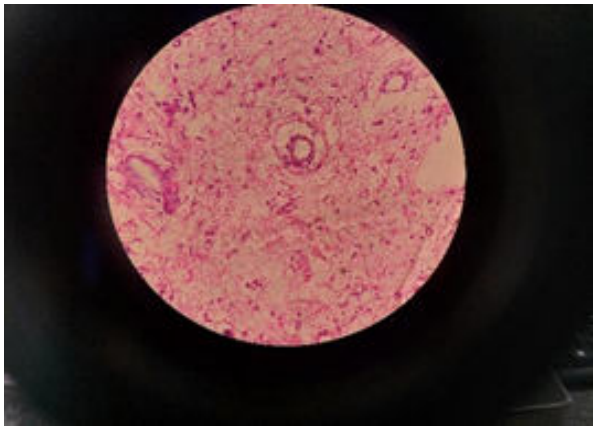

Fig 6: Another cross section of the slide showing spindle shaped cells with bipolar eosinophilic short processes (stellate cells). Dilated blood vessel with changes of hyalinisation is seen. No Evidence of mitosis/atypia/necrosis.

DISCUSSION-

Aggressive angiomyxoma is a rare, slow-growing but locally invasive, uncapsulated, estrogen-dependent myxoid neoplasm. It occurs in genital, perineal and pelvic regions of adult women, mostly in their third decade of life, the peak incidence being 31 to 35 years. [3]

Characteristic feature of this tumor is its frequent recurrence and local aggression. Unlike a superficial angiomyxoma which is usually confined to surface of the skin and is commonly seen over the head and neck region, an aggressive Angiomyxoma tends to grow deeper into the tissue and occurs exclusively in the pelvic and perineal region. [4]

About one fourth of these tumors are pedunculated. The pathogenesis of the tumor, however, still remains unclear. The lesions are composed of spindled to stellate stromal cells with thick-walled vessels of various caliber, and a myxoid extracellular matrix. A hormonal role is hypothesized in the pathogenesis of the tumor as the cells of aggressive angiomyxoma are shown to contain estrogen and progesterone receptors. Desmin immunoreactivity may be observed in the stellate stromal cells in aggressive angiomyxoma. [5]

Cytogenetic analysis and fluorescent in situ hybridization have confirmed the presence of HMGA2 gene rearrangement in aggressive angiomyxomas and have shown that they, or at least a subset of them, have the same molecular genetic background as other common mesenchymal tumors. These molecular data have led to the investigation of the role of HMGA2 immunohistochemistry in mesenchymal tumors. [7]

Being a rare entity, the diagnosis of an Aggressive Vulval Angiomyxoma may pose a dilemma to the clinician. It may be misconstrued to be a Lipofibroma, Bartholin cyst or a Labial cyst. Angiomyofibroblastoma, Fibroepithelial stromal polyps, Myxoid leiomyomas, Myxoid lipomas, Cellular angiofibroma and Smooth muscle tumors should also be taken into consideration in the differential diagnosis for a polypoidal mass arising in the perineum. [1] On rare occasions, an Aggressive Angiomyxoma metastasizes, with a few cases of multiple metastasisbeing reported in literature [6]

Wide surgical excision is the treatment of choice. Unfortunately, due to the aggressive nature of the tumor, there are documented incidences of recurrences, especially following suboptimal resection. The incidence of local recurrence is 36 to 72%. Treatment modalities like angiographic embolization of the mass, hormonal therapy with Tamoxifen, Raloxifen, Gonadotropin-releasing hormone agonist (GnRH-A) may be required cases of recurrence but they have been used with limited success. [3]

CONCLUSION:

Aggressive Angiomyxomas are a rare form of vulval masses which is slow and insidiously growing, and painless in nature. Since vulval masses have many differentials, it causes difficulty in diagnosing the lesion. Hence, a high level of clinical suspicion is needed to accurately make the clinical diagnosis, which is confirmed on histopathological examination. Surgical resection remains the first line treatment for these tumors. However, due to its locally aggressive nature, these tumors have a high recurrence rate. Hence routine long term follow up should be advised to the patient.

This unusual case of a vulval mass presenting as an aggressive vulval angiomyxoma is reported due to its rarity of occurrence.

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